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Dermatomyositis - the impact of quick diagnosis and treatment on the development of the disease. A review of current research

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ABSTRACT

Introduction: Dermatomyositis (DM) is an inflammatory disease classified among autoimmune diseases affecting young and older individuals. Due to its non-specific symptoms and insidious onset, patients and primary care physicians often overlook it. Delayed diagnosis postpones the initiation of effective treatment and causes irreversible damage to the body. *Aim of the study:* This study reviews the current literature on the clinical presentation, diagnosis, and treatment of dermatomyositis. It also summarises existing knowledge in this field. *Summary of knowledge:* Diagnosis of DM relies on clinical symptoms, including muscle weakness and skin lesions. Decreased strength in the shoulder and hip muscles hinders basic tasks like combing hair or climbing stairs. Late diagnosis can lead to severe musculoskeletal issues. Optimizing treatment outcomes involves tailoring the regimen to the lesion type, muscle involvement, specific autoantibodies, and the patient's age. *Conclusion:* Early diagnosis and timely initiation of treatment are crucial for managing dermatomyositis and enhancing the quality of life for patients. Despite the availability of various treatments, achieving clinical remission remains challenging for many patients. This highlights the need for continued research to develop more effective therapies and strategies for managing this complex disease.

Keywords: Dermatomyositis; Idiopathic Inflammatory Myopathy; Motor activity impact; Symmetrical muscle weakness

1. INTRODUCTION

Dermatomyositis is an inflammatory disease that belongs to a group of rare autoimmune diseases. The illness exhibits polymorphous cutaneous features and variable muscle involvement (Didona et al., 2020). In 1975, researchers established the initial classifications of myositis within the context of collagenoses and rheumatic diseases. The defining criteria were published in 2017 by the European League Against Rheumatism (EULAR), and the American College of Rheumatology (ACR) constituted another standard (Bohan and Peter, 1975; Lundberg et al., 2017).

Decreased strength in the shoulder and hip muscles hinders basic tasks like combing hair or climbing stairs. Late diagnosis can lead to severe musculoskeletal issues. Optimizing treatment outcomes involves tailoring the regimen to the lesion type, muscle involvement, specific autoantibodies, and the patient's age. Biological therapies offer promise in DM management. While many aspects of the disease remain incompletely understood, various topical and systemic treatments are available to manage disease activity and achieve clinical remission (Schlecht et al., 2020; Dalakas et al., 1993).

2. METHODOLOGY

Articles related to the topic were analyzed. We used the PubMed database, Google Scholar, and ScienceDirect. We based the eligibility criteria on the publication date, compliance with the topic, and the keywords. We mainly excluded articles older than ten years. We searched for English phrases including: 'dermatomyositis', 'idiopathic inflammatory myopathy', 'dermatomyositis treatment', and 'dermatomyositis epidemiology'. The authors reviewed the articles and presented collected data in the text.

3. RESULTS AND DISCUSSION

Epidemiology and Pathogenesis

Among adults, the reported incidence of the condition varies from 1.2 to 17 new cases per 1,000,000 individuals, with a prevalence ranging between 5 and 11 cases per 100,000 individuals (Sena et al., 2018). Onset typically occurs between the ages of 5 and 15 years and again between 45 and 65 years. Women are affected at approximately twice the rate of men (Meyer et al., 2015). Interestingly, there's a notable increase in the relative prevalence of DM with increasing geographical latitude from northern Europe to southern Europe (Hengstman et al., 2008). While all ethnic groups can be affected, it is more prevalent among African Americans (Findlay et al., 2015). The etiology of DM remains incompletely understood. Researchers hypothesize that autoimmune mechanisms, influenced by genetic predisposition, environmental triggers, and immune system responses, play a significant role in its development (DeWane et al., 2020).

For example, certain genetic markers, such as HLA DRB10301 and DQA10501 in Caucasians, and HLA-B7 in Asians, have been linked to DM (Thompson et al., 2018). Scientists have also associated certain medications, like immune checkpoint inhibitors, aromatase inhibitors, and statins, with the onset of DM (Yamaguchi et al., 2021; Fania et al., 2017; Schlecht et al., 2020). There have been reports of transient myositis following infections with coxsackievirus, parvovirus B19, echovirus, and influenza viruses. However, a direct causal link between viral infections and DM has not been established (Christensen et al., 1986; Leff et al., 1992). Finally, there is a hypothesis that SARS-CoV-2 infection may also be a predisposing factor for DM (Holzer et al., 2022)

Clinical Appearance and Health Implications

We can categorize the manifestations of dermatomyositis into four groups: General symptoms, skin symptoms, muscle symptoms, and organ symptoms. General symptoms include weakness, increased body temperature, and weight loss. Skin eruptions play a pivotal role in diagnosing DM. They may manifest before or concurrently with myositis, persisting even after myositis resolution (homeopathic DM). Periorbital edemas involving the bridge of the nose and eyelids are typical in the acute phase, accompanied by symmetric erythemas with a pinkish-purple or lilac hue (heliotropic erythema). Facial erythema, ranging from mild to resembling lupus erythematosus (butterfly erythema), can also occur on the cleavage (V sign), back of the neck (shawl sign), extensor sides of limbs, and backs of hands (Chansky et al., 2018). Detecting erythematous skin signs is challenging in individuals with dark skin, but chronic damage signs, such as hyperpigmentation, hypopigmentation, telangiectasia, or epidermal atrophy, are often more apparent.

Chronic damage is frequently described as "red on white" or "mottled" (Chansky et al., 2018; Marvi et al., 2012). Gottron papules, flat papules over proximal interphalangeal and metacarpophalangeal joints, or sometimes elbows, ankles, or knees (Gottron sign) are pathognomonic skin features. Periungual telangiectasia may occur with painful nail fold induration (Keining sign) (Sander et al., 2010). Other signs include increased photosensitivity, non-scarring alopecia, severe pruritus leading to lichenification from scratching, and less commonly, vesiculobullous lesions, panniculitis, skin calcinosis or flagellate-like erythema (Shirani et al., 2004). Vasculopathy may manifest as Raynaud's phenomenon or livedo racemosa. Linear hyperkeratoses and bumps may appear on the hands, ulnar aspect of thumbs, and radial aspect of second and third fingers (mechanic's hands) or on the feet (hiker's feet). They are described as very painful (Cox et al., 2017).

Researchers developed the Cutaneous Disease Area Severity Index (CDASI) to document skin involvement in dermatomyositis. It evaluates disease activity by describing inflammatory skin lesions and residual skin damage in fifteen locations, including the periorbital region, cleavage, and back of the neck (Anyanwu et al., 2015). Muscle weakness, particularly affecting proximal muscles, can develop rapidly (within days) or gradually (over weeks to months). Weakness makes tasks like getting up from sitting, climbing stairs, and lifting heavy objects challenging and may also cause discomfort. In patients with the acute onset of the disease, getting up from a lying position is difficult. Muscle weakness is accompanied by pain and soreness when pressed. Over time, muscle weakness may progress to involve muscles used for swallowing, breathing, and those in the back of the neck. In some cases, the heart and lungs may also be affected.

Amyopathic DM, comprising 5-20% of cases, is characterized by typical skin manifestations without any muscular involvement, either clinically or detectable through laboratory tests. If a patient has had amyopathic DM for two years, it's unlikely that muscle involvement will develop after that (Sunderkötter et al., 2016; Euwer and Sontheimer, 1991). This form of DM is more prevalent in light-skinned Europeans than in Black Americans (Chansky et al., 2018). To accurately evaluate the severity and progression of myositis, use a validated method such as Manual Muscle Testing 8 (MMT-8). MMT-8 assesses the strength and function of eight different muscles or muscle groups (Rider et al., 2010). It's important to note that DM is not associated with sensory loss, ptosis, involvement of the extraocular muscles, or abnormal reflexes, which can aid in distinguishing it from other neuromuscular disorders (Findlay et al., 2015).

Dermatomyositis frequently affects the lungs, leading to interstitial lung disease (ILD), often overlooked or detected late (Sunderkötter et al., 2016; Marvi et al., 2012). Lung involvement typically presents with symptoms like exertional dyspnea, cough, and decreased exercise tolerance, usually developing after muscle involvement. However, subclinical courses, especially in the early stages of the disease, are also possible (Didona et al., 2020; Callen, 2000; Dalakas and Hohlfeld, 2003). "Antisynthetase syndrome" is marked by three main features: Muscle inflammation, joint inflammation, and lung disease affecting the tissue between the air sacs. Additionally, pericarditis, dilated cardiomyopathy, or coronary heart disease with associated ECG alterations may occur, potentially leading to heart failure (Marvi et al., 2012). Reports indicate subclinical cardiac involvement in up to 50% of patients, with myocarditis occurring in about 30% of cases (Didona et al., 2020; Callen, 2000; Dalakas and Hohlfeld, 2003; Zhang et al., 2012).

Gastrointestinal manifestations such as dysphagia, loss of appetite, constipation, and occasionally diarrhea can result from edemas and muscle weakness in the gastrointestinal tract. Esophageal involvement in DM patients typically manifests as dysphagia due to loss of pharyngo-esophageal muscle tone, increasing the risk of aspiration pneumonia (Didona et al., 2020; Callen, 2000; Dalakas and Hohlfeld, 2003). Joint symptoms resembling those of rheumatological diseases may also occur in DM, making rheumatological disease an essential consideration for differential diagnosis and potential misdiagnosis. Organ involvement symptoms may precede skin or muscle changes (Cottin et al., 2003). Statistical surveys link about 30% of dermatomyositis cases to malignancy (Hill et al., 2001). Detection of anti-TIF-1γ antibodies and anti-NXP-2 antibodies, in particular, suggests a high likelihood of paraneoplastic DM (Hoshino et al., 2010; Fiorentino et al., 2013).

However, tumor screening is recommended for all forms of DM (Cobos et al., 2020). Clinical signs of paraneoplastic dermatomyositis include skin ulceration, mouth enanthema, pronounced skin and muscle symptoms, acute disease onset, and involvement of pharyngeal, diaphragm, and distal limb muscles (András et al., 2008; Fardet et al., 2009). In most cases, doctors diagnose the associated malignancy within one year after the onset of myositis. However, a tumor may already be present at the time of myositis diagnosis or even before symptoms manifest. The underlying malignancies often include adenocarcinomas of the lungs, cervix, ovaries, breasts, pancreas, bladder, or stomach (Chen et al., 2010).

From the First Symptoms to Diagnosis

During medical interviews, in addition to asking general questions about the onset and progression of symptoms, it's crucial to inquire about any difficulties in daily activities. The pathognomonic symptoms of dermatomyositis include heliotropic erythema, Gottron's papules, and Gottron's sign. In a histopathological examination of skin biopsy material, it can be observed degeneration of the basement membrane zone with vacuolized basal keratinocytes, subepidermal lymphocytic infiltration (interface dermatitis), epidermal atrophy, and interstitial mucin deposits in the dermis. Histologically, DM closely resembles lupus erythematosus, making differentiation challenging (Sunderkötter et al., 2016). In laboratory tests, serum creatine kinase (CK) levels typically rise to 10–50 times the average value. CK serves as a marker for assessing current muscle damage. Increased AST levels are typical in DM, as they are in other forms of muscle damage, while LDH may show a non-specific increase.

Laboratory investigations should include a comprehensive, reliable evaluation of antibodies tailored to the symptoms (Pfleiderer et al., 2004). MRI and electromyography (EMG) are essential diagnostic tools. Muscle ultrasound is also potentially useful. Muscle MRI helps in selecting the optimal site for muscle biopsy and aids in diagnostic classification, prognostic evaluation, and predicting treatment responses. If T1-weighted images demonstrate significant fatty transformation in weak muscle groups, immunosuppressive treatment may be ineffective. T2-weighted imaging can reveal muscle edema resulting from acute inflammation. EMG typically reveals spontaneous discharges and polyphasic potentials. The primary method to confirm myositis and distinguish it from other neuromuscular diseases is a muscle biopsy.

Perimysial infiltration, increased primary histocompatibility complex class I (MHC-I) expression, complement deposition on capillaries and the sarcolemma, perifascicular atrophy, and muscle fiber necrosis are usually present on histopathological examination of the muscle. When the clinical presentation of DM is clear, with typical skin manifestations, objectively confirmed muscle weakness, and significantly elevated CK levels, a muscle biopsy may not be necessary. For patients diagnosed with amyopathic DM (a diagnosis of exclusion), clinical and laboratory assessments to detect muscle involvement should be conducted over two years (Tomasová-Studynková et al., 2007). The European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) have jointly developed and formally accepted new criteria for classifying dermatomyositis.

These criteria achieve a sensitivity of 93% with a biopsy and 87% without a biopsy, with a specificity of 88% with a biopsy and 82% without a biopsy (Pinto et al., 2019). The criteria should be used to distinguish idiopathic myositis from other systemic connective tissue diseases. All patients with DM should undergo pulmonary function tests, including an assessment of CO2 diffusion capacity, due to the possibility of subclinical DM. Electrocardiography (ECG) helps detect early signs of cardiac involvement, such as arrhythmias, conduction abnormalities, repolarization abnormalities, and signs of left ventricular and left atrial hypertrophy. ECG abnormalities are present in about one-fourth of patients. Dysphagia can be assessed through esophageal manometry. Furthermore, endoscopy can examine related issues, such as dysmotility, erosions, and ulcers (Lundberg, 2006).

Autoantibodies as Valuable Biomarkers

Autoantibodies are valuable biomarkers for predicting prognosis and systemic involvement in dermatomyositis patients. However, not all autoantibodies are specific to DM and may also be found in other diseases (Waldman et al., 2020). Myositis-specific antibodies (MSA) play a role in various cellular processes, including DNA repair, transcription, protein translation, and recognition of viral structures. In dermatomyositis, most patients have only one type of MSA (Wolstencroft and Fiorentino, 2018). The most known MSA with their targets, frequency, and clinical phenotypes are in Table 1 (Wolstencroft and Fiorentino, 2018; Biddle et al., 2022; Hou et al., 2019; Hodgkinson et al., 2021; Rogers et al., 2017; Fiorentino et al., 2015; Bernet et al., 2016; Fujimoto et al., 2016; Satoh et al., 2017; Galindo-Feria et al., 2022).

Differentiate the skin symptoms of dermatomyositis primarily from rosacea, seborrheic dermatitis, and contact dermatitis. Lupus erythematosus is also a pertinent alternative diagnosis due to the potential UV association of lesions, similar clinical appearance (particularly in later stages), and typically challenging histopathological differentiation. Consider possible causes for myositis, including infectious agents, genetic predispositions, pharmaceuticals or toxins, and metabolic or endocrine disorders (Didona et al., 2020; Day and Limaye, 2019).

Table 1 Myositis-specific antibodies (Zhang et al., 2012) and their target antigens (Galindo-Feria et al., 2022). Percentages are approximate values.

MSA: Autoantibodies against	Frequency in the adult form of DM (caucasian patients)	Clinical associations
MDA-5	30%	High risk of developing rapidly progressive ILD
Mi-2	20% -30%	Low risk of ILD and malignancy
NXP-2	5%	Milder disease course, recurrent muscle weakness, and severe dysphagia
TIF-1γ	14-2%	Severe cutaneous involvement, oval, partly eroded, ulcerating lesion on the patient's palate
SAE 1/2	5-8%	Weight loss and elevated inflammatory markers, mild ILD, and malignancy
Jo-1	5-20%	Classic DM, ILD, Antisynthetase syndrome

Treatment and its effectiveness

The time that passes from the onset of the first symptoms to the actual onset of treatment is a critical factor that influences the further prospects of the patient. Each week of delay in proper therapy can have serious consequences consequences. The 5-year mortality rate for patients with DM was approximately 50% before the advent of glucocorticoids for treatment (Lundberg, 2006). However, early detection and immunosuppressive therapy can prevent rapid disease progression. Therefore, despite the availability of steroids, treatment remains challenging. Before initiating treatment, assess several factors, such as disease activity, age, and concurrent medical conditions (Schlecht et al., 2020). While most patients initially respond well to immunosuppressive therapy, many struggle to control inflammatory activity in the muscles over time, leading to progressive muscle weakness (Sunderkötter et al., 2016).

Immunosuppression with corticosteroids remains the cornerstone of therapy. Patients with localized skin lesions should receive treatment with topical class II or IV steroids. Topical treatment for erythema and pruritus is usually sufficient, although lesions on the head and neck are often refractory (Schlecht et al., 2020). Systemic corticosteroids are the primary treatment in patients with myositis. However, their efficacy as monotherapy is reduced in patients with clinically amyopathic dermatomyositis (Iorizzo and Jorizzo, 2008). It is necessary to regularly monitor side effects and contraindications to the use of high-dose glucocorticoid therapy, such as hypertension or decompensation in diabetes (Van-de-Vlekkert et al., 2010). Recommend combination treatment with steroids and immunosuppressive medicines such as methotrexate (MTX), azathioprine (AZA), or mycophenolate mofetil (MMF) for moderate and severe DM.

Steroid monotherapy is only appropriate in very mild cases. Randomized controlled trials did not show a statistically significant difference in muscle strength one-year post-treatment between patients receiving a combination of methotrexate and prednisolone compared to those treated with prednisolone alone (Ibrahim et al., 2015). However, in a recent series involving 13 dermatomyositis patients, complete resolution was observed in 30% of cases, with another 30% experiencing near-complete resolution (Kasteler and Callen, 1997). AZA showed positive results in patients with esophageal involvement and similar efficacy to methotrexate in treating patients with polymyositis and DM with muscle involvement (Joffe et al., 1993).

A retrospective open-label review demonstrated improvement in 12 patients with classical dermatomyositis regarding cutaneous and muscle features within eight weeks of initiating mycophenolate mofetil (Edge et al., 2006). With proper treatment of potential side effects, the first therapeutic effects of AZA, MMF, and MTX become visible after a few weeks. An alternative to the above therapies may be cyclosporine (CsA) and intravenous immunoglobulins (IVIg), alone or in combination. IVIg is also suitable for pregnant or breastfeeding patients (Dalakas et al., 1993). Further therapeutic escalation may involve rituximab (RTX) or cyclophosphamide (CYC) treatment. In a significant randomized trial, 83% of DM patients who had previously shown no clinical improvement with systemic corticosteroids and at least one steroid-sparing agent experienced improvement after rituximab treatment.

Furthermore, RTX affects nail fold capillary abnormalities, inverse Gottron's papules, and digital ulcerations. The approach enabled a quicker reduction of systemic corticosteroids (Oddis et al., 2013). Cyclophosphamide has demonstrated effectiveness in treating

severe interstitial lung disease. It is considered for patients with severe systemic DM who do not respond to initial and secondary treatments, as well as in cases of severe antisynthetase syndrome. Intravenous immunoglobulin can serve as a steroid-sparing agent in patients with refractory muscle and skin manifestations, typically prescribed for those who do not respond to antimalarials and methotrexate. According to a retrospective study, 83% of patients experienced significant improvement in their cutaneous symptoms, irrespective of the subtype of dermatomyositis (Lam and Vleugels, 2012).

Numerous reports have highlighted the positive impact of IVIG on ulcerations and vasculopathy lesions in individuals with dermatomyositis. Research demonstrates that moderate physical therapy is advantageous in managing dermatomyositis. Endurance training, combined with resistance training, is paramount (Van-Thillo et al., 2019). Patients experiencing mild gastrointestinal involvement may find relief from bulking agents such as linseed or macrogol. It's noteworthy that skin symptoms in dermatomyositis may not consistently respond well to systemic immunotherapy, even after myositis symptoms have subsided. Table 2 presents data comparing the quality of life of patients undergoing treatment and those not receiving medication for dermatomyositis.

The impact of treatment on the development of the disease

Dermatomyositis (DM) is a multifaceted autoimmune disorder that can significantly diminish patients' quality of life. The comparison of quality of life, symptoms, and severity in patients with dermatomyositis who receive treatment versus those who do not reveal significant differences in patient outcomes. The studies' results provide a comprehensive understanding of the impact of therapy on DM. Patients receiving treatment for DM experience a marked improvement in their quality of life. Robinson et al., (2015) demonstrated significant improvements in Skindex-29 scores, which measure various aspects of quality of life including emotions, functioning, and symptoms, among treated patients. The study found that responders to treatment showed a decrease in these scores, indicating better emotional well-being, improved functionality, and fewer symptoms. In contrast, untreated patients suffer from a poorer quality of life as they continue to endure severe symptoms and associated emotional distress.

Reducing symptoms, particularly itch and pain, is another significant advantage of therapy. Robinson et al., (2015) reported that treated patients experienced a substantial decrease in itch and pain scores on the Visual Analogue Scale (VAS). The responders' median itch score decreased from 2.4 to 0.4, and the pain score from 2.4 to 0.8, whereas nonresponders saw no such improvement and, in some cases, even experienced worsening symptoms. This stark contrast underscores the importance of effective treatment in managing the debilitating symptoms of DM. The Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) scores also reflect the positive impact of treatment. Robinson et al., (2015) found that responders experienced a significant decrease in CDASI activity scores, dropping from 24 to 9. This suggests a notable reduction in skin disease activity.

Table 2 Quality of Life, Symptoms, and Severity in Dermatomyositis (Robinson et al., 2015; Kim et al., 2009).

Aspect	Treated Patients	Untreated Patients
Quality of Life	Improved significantly with treatment, particularly regarding emotional well-being, functionality, and symptom relief.	Poor quality of life due to untreated symptoms leads to significant morbidity.
Symptoms (Itch and Pain)	Significant reduction in itch and pain scores with treatment.	Persistent and often severe itch and pain without treatment.
Skin Disease Activity	Marked improvement in cutaneous disease activity (measured by CDASI) with appropriate treatment.	High skin disease activity remains without treatment, contributing to poor quality of life.
Muscle Strength and Function	Treated patients show improved muscle strength and function over time, often achieving remission with aggressive treatment protocols.	Muscle weakness and dysfunction persist, leading to increased disability and decreased daily functioning.
Disease Severity	Treatment reduces overall disease severity, including systemic involvement and complications like calcinosis.	Higher severity with untreated systemic involvement and complications such as interstitial lung disease and malignancy

		risks.
Psychosocial Impact	Emotional and social well-being significantly improved with effective management.	Untreated patients often experience social isolation, depression, and anxiety due to visible skin symptoms and physical limitations.
Mortality Risk	Comprehensive treatment strategies reduce the risk of mortality.	Increased risk of mortality associated with untreated disease progression and complications.

Nonresponders did not exhibit such improvements, indicating persistent and severe skin involvement in untreated DM. Kim et al., (2009) also emphasized that aggressive treatment protocols improve muscle strength and function. Untreated patients continued to struggle with muscle weakness and dysfunction, significantly impacting their daily activities and overall mobility. Treatment substantially reduces overall disease severity and the risk of complications. Kim et al., (2009) reported that aggressive treatment protocols not only improved disease outcomes but also reduced the occurrence of calcinosis and other complications. In untreated patients, the severity of DM remains high, leading to more frequent and severe complications such as interstitial lung disease and increased malignancy risks.

The psychosocial impact of DM is profound, with untreated patients often facing significant emotional and social challenges. Robinson et al., (2015) found that effective treatment considerably improved the emotional and social well-being of patients, reducing feelings of depression and social isolation caused by visible skin symptoms and physical limitations. Untreated patients are more likely to suffer from these psychosocial burdens, further deteriorating their quality of life. The probability of mortality is another critical aspect where treatment plays a pivotal role. Aggressive and timely intervention has been shown to lower mortality rates in DM patients by managing symptoms and preventing severe complications. Untreated patients face a higher mortality risk due to the progression of the disease and its associated complications (Robinson et al., 2015; Kim et al., 2009).

4. CONCLUSION

The evidence indicates that treatment for dermatomyositis significantly improves patients' quality of life, reduces symptoms and disease severity, and decreases the risk of complications and mortality. These findings underscore the importance of early and aggressive treatment in managing DM effectively.

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Conflict of interest
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Data and materials availability
All data sets collected during this study are available upon reasonable request from the corresponding author.

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